Cardiac Sarcoidosis versus Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

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Patients with cardiac sarcoidosis may present with clinical and morphological features similar to ARVC/D or cardiomyopathy (Ott 2003). Sarcoidosis is an inflammatory granulomatosis entity of unknows cause, characterized by multisystemic involvement. Practically no organ is immune to sarcoidosis; most commonly, in up to 90% of patients, it affects the lungs. (**Hoitsma 2004**). The most commonly involved organ in sarcoid related death has been reported to be the lung in western countries, while it was the heart in the Japanese autopsy series. (**Iwai 1994**).

The diagnosis of myocardial sarcoidosis is difficult and frustrating. Its clinical manifestations depend on the location and extent of granulomatous inflammation, and the symptoms and signs range among benign arrhythmias, heart block, intractable CHF, intense chest pain, to fatal VF. (Sharma 2003).

The ECG finding may be normal or may reflect every degree of block of the atrioventricular junction and bundle of His and every type of arrhythmia along with nonspecific ST-T-wave changes.

Cardiac sarcoidosis should be considered in all young patients with unexplained conduction disorders,(**Kollermann 2001**) CHF or in cases of SCD (**Lip 1996**).

In extensive forms are frequently pseudo myocardial infarction patterns with pathological Q waves on ECG. (**Shindo 1998**).

MRI abnormalities, consisting of cardiac signal intensity and thickness, with the following three patterns:

1. Nodular;
2. Focal increase in signal on gadolinium diethylenetriamine Penta acetic acid-enhanced, T1-weighted images;
3. Focal increased signal on T2-weighted images without gadolinium uptake.

The improvement or stability of the MRI findings is correlated with clinical features.

With corticosterotherapy, the MRI images improved either partially or completely, whereas.

The cardiac MRI may find its usefulness as a guide to obtaining EMB specimens and to monitoring the response of the disease to treatment.

The study is small and lacks a correlation of myocardial histology with MRI features. However, the study clearly calls for a large multicenter trial.

The most significant drawback of MRI is that the patient with a pacemaker and/or automatic ICD will not be able to take advantage

of it. In such patients, 201Tl scanning remains the test for assessing myocardial damage.

Cardiac PET using (18) F-FDG under fasting conditions (fasting (18) F-FDG PET) is a promising technique for identification of cardiac sarcoidosis and assessment of disease activity. The methodology can detect the early stage of cardiac sarcoidosis, in which fewer perfusion abnormalities and high inflammatory activity are noted, before advanced myocardial impairment. The sensitivity of fasting (18) F-FDG PET in detecting cardiac sarcoidosis was 100%, significantly higher than that of (99m)Tc-MIBI SPECT (63.6%) or (67)Ga scintigraphy (36.3%). The accuracy of fasting (18) F-FDG PET was significantly higher than (67) Ga scintigraphy. (**Okumura 2004**.).

An EMB is preferable, but the procedure has sensitivity as low as 20% (**Uemura, 1999**). Others author referred sensitivity approximately of 50% thus, the search for a safe, reliable, and easily available diagnostic test for cardiac sarcoidosis continues. The pathological feature is the presence of noncaseating granulomas that eventually form fibrotic scars. Table 1

The table 1 shows the principal differences between the two entities:

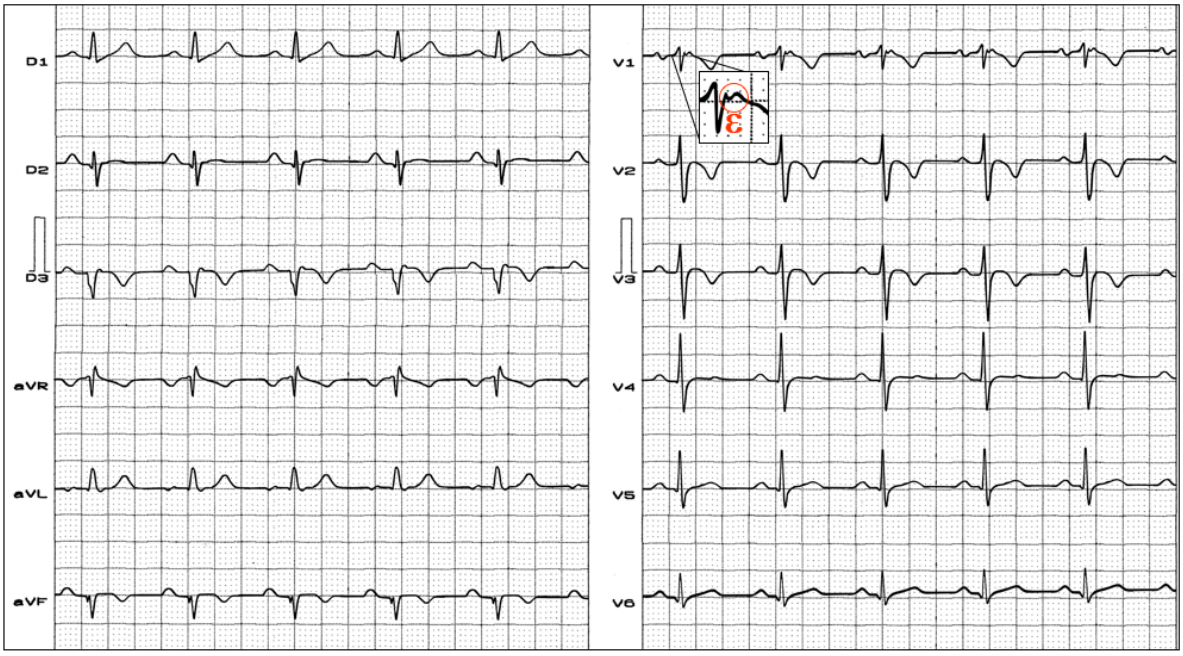
	Cardiac Sarcoidosis	ARVC/D
Family history:	Absent.	Present in 30% to 50% of cases. When the disease is identified genetic screening should be performed among patient's family members.
Gender (M/F):	1 to 1.	2.9 to 1
Age at presentation:	Young or middle-aged adults.	Adolescents and young adults, perhaps There are rare references in childhood
Multisystemic involvement:	Yes.	No.
Chest pain:	Intense chest pain is referred.	No.
Clinical myocardial restrictive features:	Possible.	No.

Mitral regurgitation:	Is common.	Only in late stage with involvement of LV.
Pseudo myocardial infarction patterns on ECG:	Frequent in extensive forms.	No.
Epsilon wave on ECG	Rarely observed (figure 1)	Mayor depolarization criteria, but not pathognomy
Chest roentgenogram:	Bilateral hilar lymphadenopathy.	Eventually RV cardiomegaly.
Lungs affectation:	In up to 90% of patients. Cor pulmonale is frequent.	No.
Pathological features:	Noncaseating granulomas that eventually form fibrotic scars.	Typical fibro-fatty replacement of the RV myocardium on dysplasia triangle.
More comum cardiac sites involved:	LV free wall and interventricular septum.	RVOT, RVIT, and apex of RV.
Pericardial effusion:	Are not uncommon.	Absent.

Improved MRI images with corticosteroids:	Yes.	No.
Therapy with corticosteroids, hydroxychloroquine, methrotexate or cyclophosphamide:	Sometime are indicated. (**Mitchell 1997**). Immunosuppressive and antcytokine treatments can be effective in severe systemic sarcoidosis and should be considered in sight-threatening disease.	No.

Figure 1

Typical ECG of ARVC/D with epsilon wave



Clinical diagnosis: cardiac sarcoidosis.

ECG diagnosis: SÂQRS -60º, negative T wave from V1 to V3, Epsilon wave (ε) in V1.

Characteristics of epsilon or Fontaine wave in ARVC/D

Intrinsic features: they are small notches or oscillations in variable quantities (1, 2, 3 or more).

Location: at the end of QRS in the J point or onset of ST segment (there is no consensus about this).

Leads: observed in right precordial leads; however Dr. Li Zhang et al, found the ε wave in the leads of the frontal plane, especially in inferior leads.

Frequency in ARVD: approximately 15-30% of cases in 12-lead ECG. This percentage increases if we use the ECG with the modified protocol.

Value of criterion: considered to be a major criterion for diagnosis by the Task Force for ARVC/D diagnosis (**McKenna 1994; Fontaine 1999**).

High resolution ECG: observed more frequently with this method.

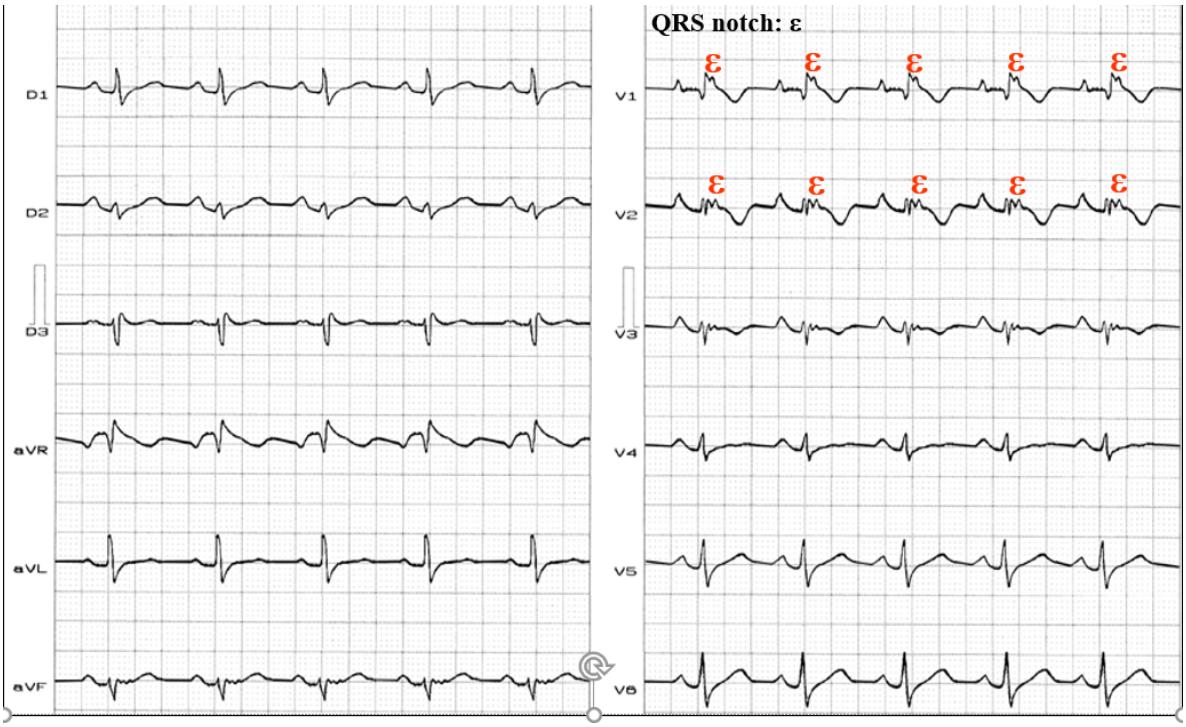
Pathognomonic character: in spite of the characteristics in ARVC/D, they are not pathognomonic, since they have been described in other diseases associated with myocardial damage: RV infarction, inferior or dorsal (**Zorio 2005**), sarcoidosis **(Santucci 2004**), Brugada syndrome, sickle cell anemia (**Hurst 1998**), etc.

Meaning: late posterior potentials (PP) that occur in the RV free wall in patients with ARVC/D.

Inversion of T wave in leads V1-V3 and/or ε wave found in 70% of patients with ARVC/D. Epicardial electrophysiological studies in dysplastic areas reveal the LP that occur at the end of the QRS complex, in the J point, and at the onset of the ST segment, are explained by fibro-fatty substitution of myocardial tissue **(Fontaine 1984**). Figure 2

Figure 2

Typical ECG of ARVC/D



Sinus rhythm, CRBBB, terminal notch located in the J point (epsilon wave). The epsilon wave could be the result of delayed activation in the RV. It is visible from V1 to V3 and in the frontal plane leads. T wave inversion is observed in V1 to V3, characteristic of ARVC/D.

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